Comment. Dyskeratosis congenita is a multisystemic congenital syndrome classically typified by reticulate hyperpigmentation of the skin, dystrophy of the nails, and leukoplakia of the mucous membranes. A host of other abnormalities may be associated, including pancytopenia.1

Exclusive of the intracranial calcifications, our patient's constellation of abnormalities, including the retinopathy1 and hypoplastic anemia, clearly fits into the spectrum of dyskeratosis congenita. The intracranial abnormalities are not consistent with the watershed infarcts typically seen with perinatal asphyxia, nor were they consistent with any other known neurocutaneous syndrome. The lesions are most consistent with in utero infection, but our patient had no serologic nor other clinical evidence to support such a contention. Metabolic disease with dystrophic calcification can be ruled out on clinical and laboratory grounds. Normal long bone roentgenograms ruled out osteopetrosis. Thus, we assume the calcifications are due to dyskeratosis congenita and account for her global developmental deficits.

We echo the comments made previously by Mills et al.1 and Lieblich et al.1 that present and future cases of dyskeratosis congenita be studied and followed for the presence of intracranial calcifications and associated neurologic or developmental defects.

Patricia A. Duprey, M.D., and James W. Steger, M.D.
Naval Hospital, San Diego, CA 92134-5000

REFERENCES


Herpes simplex virus infections and cimetidine therapy

To the Editor: We read the case report by Westheim et al. (J Am Acad Dermatol 1987;17:875-80), concerning an acyclovir-resistant herpes simplex virus infection in an immunocompromised host, with interest and sympathy. Briefly, their patient had chronic lymphocytic leukemia and a progressive chronic mucocutaneous herpes simplex virus infection involving the mouth and face for a duration of approximately 84 and 4 months, respectively. Neither intravenous acyclovir nor vidarabine treatment was effective.

Recently, we reported rapid improvement in severe mucocutaneous herpes simplex virus infections in several profoundly immunocompromised patients treated with cimetidine (300 mg administered orally or intravenously every 6 hours).1,3 Their diagnoses included the acquired immunodeficiency syndrome (AIDS), leukemia, and gastric carcinoma. Similar improvements in extensive herpes simplex virus infections in cancer patients treated with oral cimetidine have also been reported previously by Van Der Spuy et al.5

Cimetidine is a competitive antagonist of the H2 histamine receptor. It is commonly used to treat peptic ulcer disease and has a low frequency of side effects and adverse reactions. Since histamine-responsive H2 receptors are present on a subclass of suppressor T lymphocytes, cimetidine may also function as an immune modulator. An augmented cell-mediated immune response might result from cimetidine interfering with the stimulation of these cells by blocking their receptors from histamine.4 The pharmacophysiology for the clinical response observed in several immunocompromised patients with progressive herpes simplex virus infections to cimetidine is presently undefined. A direct antiviral effect or an indirect effect resulting from the augmentation of the host's cell-mediated immunity is a possible mechanism.

The acquisition, severity, development and maintenance of latency, and recurrence frequency of herpes simplex virus infection are influenced by the immune response of the host. The infections are usually more severe and extensive in immunocompromised patients, especially those with defects in cell-mediated immunity. Acyclovir is an effective drug for the treatment of most herpes simplex virus infections in both immunocompetent and immunosuppressed patients. The emergence of acyclovir-resistant herpes simplex virus infections has prompted the evaluation of other antiviral agents.6 A compound that is unrelated pharmacologically to acyclovir could be especially useful in these patients. This consideration, coupled with the previous reports of healing of herpes simplex virus infections in immunosuppressed hosts treated with cimetidine, suggests that a randomized, double-blind trial using cimetidine may be warranted.

Philip R. Cohen, M.D.,* and Razelle Kurzrock, M.D.**
College of Physicians and Surgeons of Columbia University, New York, NY 10032,* and The University of Texas System Cancer Center, M.D. Anderson Hospital and Tumor Institute, Houston, TX 77030**
REFERENCES


Reply

To the Editor: This observation is of considerable practical importance and confirms prior reports such as that of Jorizzo et al., in which they demonstrated that cimetidine could reverse selected immunodeficiencies in four adult patients with chronic mucocutaneous candidasis, and Richardsmeier et al., in which they demonstrated restoration of delayed hypersensitivity responses while on cimetidine in patients with squamous cell carcinoma of the head and neck. I certainly agree with Drs. Cohen and Kurzrock that, since cimetidine is a relatively safe drug, and since it does indeed have an effect on restoring cellular immunity, its further use in immunocompromised patients seems warranted.

W. Mitchell Sams, Jr., M.D.
University of Alabama at Birmingham
Birmingham, AL 35294

REFERENCES


Vulvar and esophageal leiomyoma

To the Editor: In reviewing the literature on smooth muscle tumors, I uncovered an error in a JOURNAL Clinical Review article. The article stated, "Multiple leiomyomas of the skin have been reported in association with uterine leiomyomas and esophageal leiomyomas." The reference cited did list "cutaneous, uterine, and/or esophageal leiomyomas" as hereditary tumors. This was extracted from a chapter in a book by the same author, which, in turn, was drawn from another author's literature review. The original case reports did note association of hereditary cutaneous and uterine leiomyomas, but esophageal leiomyomas were associated with vulvar leiomyoma only and never with cutaneous leiomyoma.

Robert M. Rosen, D.O.
Coney Island Hospital, Brooklyn, NY 11235

REFERENCES


Reply

To the Editor: Dr. Rosen is correct. In my reading of the literature in preparation for the article, I assumed the reported association of multiple leiomyomas of the skin with "uterine and/or esophageal leiomyomas" in the table of Mulvihill and McKeen referred to their own observations from the reference given. It was, in fact a secondary source, the primary being Dr. McKusick's. This emphasizes a lesson I thought I had learned, namely, always to confirm primary sources. I thank Dr. Rosen for his correction.

J. E. Jelinek, M.D.
15 W. 12th St., New York, NY 10011

Recent urticaria book

To the Editor: I read with great interest the "Periodic Synopsis" on "Immediate Hypersensitivity" by Keahey et al. in the November 1987 issue of the JOURNAL on page 826. On perusal of the recommended books on urticaria, I was surprised not to find any mention of the recent monograph on urticaria by B. M. Czarnetzki, which was available in bookstores by early 1986. This book has so far received many good reviews in a number...